

# Concurrence of Primary Cutaneous Extra Mammary Paget's Disease and Squamous Cell Carcinoma in situ of Vulva: A Case Report

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## ABSTRACT

Extramammary Paget's disease and vulvar intraepithelial neoplasia are common lesions of vulva, but synchronous occurrence is rare in a same location. Herein we describe a concurrence of primary cutaneous extra mammary Paget's disease and squamous cell carcinoma in situ of vulva in an Iranian women. A 59 year old woman, initially presented to Kosar teaching hospital gynecology clinic April 2017 with a single, well defined, scaly, white ulcerated mass, 2 cm in diameter on right minor labia, but other examinations were normal. She had no urinary or gastrointestinal symptoms. Incisional biopsies from the mass represented concurrence extramammary Paget's disease and VIN3. She underwent radical vulvectomy. No evidence of disease recurrence was noted after 16 months follow up. Participant consent was obtained before patient was enrolled in this study. There are lot of diseases that involve genitalia and lead to vulvar lesions. But in this rare case, we diagnosed concurrence of extramammary Paget's disease and vulvar intraepithelial neoplasia in a same location. Additionally, the clinical presentation as a vulvar mass was found unusual. Therefore, we report the case to sensitize gynecologists and pathologists for uncommon pathologies and their manifestations in vulva.

**KEYWORDS:** Extramammary Paget's disease, Intraepithelial neoplasia, Vulve

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## Introduction

Paget's disease is a rare neoplasm of the nipple (1). Extramammary Paget's disease (EMPD), which was first described by Crocker in 1888 (1), is an intraepithelial adenocarcinoma (2) in apocrine cells rich glandular regions (1). It's more common in Caucasian women and Asian men (1).

The most frequently affected areas include: vulva, scrotum, penis and perineum (3). Pruritus is the most common patient's symptom (4). Standard treatment is a surgical resection with wide safe margins (2). Squamous intraepithelial lesions were first described by Bowen in 1912, it is also named squamous cell carcinoma (SCC) in situ or Bowen disease, but some years after, this term changed to vulvar intraepithelial neoplasia (VIN) (5). Associated symptoms are pruritus, pain and skin lesion (4). Lesions can be red, white or pigmented, either smooth or ridged and present as striae or ulcers. However, it often manifests by erythematous plaques and is uncommon to involve genital area (2). Surgical treatment is an effective method in removal of premalignant lesions but in VIN, relapse is very common (2). EMPD and VIN alone are common lesions of vulve, but concurrence of these neoplasms in a same lesions, is a rare event (1,5).

As mentioned before, EMPD and in situ SCC usually manifest as plaque (2), but in this case, a patient is presented with a vulvar mass, which is an uncommon presentation.

Tumoral cells of VIN3 origin from the intra-epithelial portion of the hair follicle (acrotrichium) (6) or arise from multipotential cells situated along the epidermal basal layer that differentiated along squamous cells (7). On the other hand tumoral cells of EMPD derive from the intraepithelial part of glands (acrosyringium) (8) or arise from multipotential cells situated in the direction of the epidermal basal layer that transform along sweat glands series (9). About the origin of two above lesions, the second interpretation being the more acceptable, explains concurrence of them in a lesion (8,9).

Herein we describe a concurrence of primary cutaneous extramammary Paget's disease and squamous cell carcinoma in situ of vulva, an Iranian women in order to sensitize gynecologists and pathologists to this diagnostic challenge.